Valvar stenosis in truncus arteriosus

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SUMMARY Twenty three morphological specimens of truncus arteriosus were examined for evidence of stenosis of the semilunar valve. One third showed good evidence of stenosis as judged by careful measurement of the valve orifice, the valve ring, and the maximum diameter of the truncus. Correlation with measured pressure gradients was poor, but angiography and cross sectional echocardiography were better predictors of stenosis. Stenosis was invariably associated with cusp dysplasia and was more common in valves with two or four cusps.

Stenosis of the semilunar valve in truncus arteriosus is not described in standard clinical and pathological texts of paediatric cardiology. A series of morphological reviews[1–6] totalling 251 cases of truncus arteriosus yielded only one example of "probable" valve stenosis[2] and one of suggested stenosis.[5] Two other series of 180[7] and 79[8] necropsy specimens described stenotic truncus valves in 5·6% and 11% respectively. In addition, there are several isolated case reports of truncus valve stenosis.[9–14] The first antemortem diagnosis was made by Lee *et al* on the grounds of a systolic pressure gradient across the valve at cardiac catheterisation.[11] They remarked upon the apparent rarity of the condition. A retrospective diagnosis on similar grounds was made by Burnell *et al*[10]; they too emphasised the rarity of truncus valve stenosis.

In a retrospective study we reviewed 24 necropsy specimens of truncus arteriosus with particular emphasis on the morphological evidence for stenosis of the truncus valve.

Patients and methods

Twenty three morphological specimens of truncus arteriosus were identified in the pathological collection of this hospital. The case records, including radiological, electrocardiographic, and cardiac catheterisation data, were reviewed. Particular emphasis was given to identifying evidence of stenosis of the truncus valve. M mode echocardiograms were obtained using a Smith Kline Echoline instrument, and cross sectional data using an Advanced Technol-

ogy Laboratory instrument. At cardiac catheterisation pressure measurements had been made using fluid filled catheters. Simultaneous recordings from the truncus and either ventricle, or a withdrawal recording from truncus to right ventricle, were available in 15 patients (Table).

Each morphological specimen was examined and classified, the number of the truncus valve cusps and the presence and degree of dysplasia were noted, and any associated cardiac malformations were identified. The truncus valve was viewed from above and the cusps were opened to the maximum extent possible. Calibrated calipers were used to measure the maximum diameter of the valve orifice, the valve ring, and the diameter of the truncus at its widest point (Fig. 1). In each individual the degree of stenosis was expressed according to the following formulas:

$$\text{Absolute stenosis} = \frac{(\text{ring diameter}^2 - \text{orifice diameter}^2)}{\text{valve ring diameter}^2} \times 100$$

$$\text{Relative stenosis} = \frac{(\text{truncus diameter}^2 - \text{orifice diameter}^2)}{\text{truncus diameter}^2} \times 100$$

Results

All the cases presented in cardiac failure in the neonatal period. There were 15 (65%) boys and eight (35%) girls. Their ages at death ranged from 4 days to 7 years (median 4 weeks). Precordial thrills were present in 10 (43%) and systolic murmurs in all cases. In five cases a diastolic murmur was also present. Radiologically all patients had cardiomegaly and pulmonary plethora. Electrocardiography showed biventricular hypertrophy in 12 (52%) cases, right ventricular hypertrophy in five (22%), and left ventricular hypertrophy in two (9%). There was T wave inversion in the left precordial leads in 14 (61%) cases. In only

Table *Investigative and morphological data relevant to truncus valve stenosis*

Case No	Age at death	Echocardiography*	Angiocardiography*	Pressure gradient (mm Hg)	Cusps		% Stenosis	
					No	Dysplasia†	Absolute	Relative
1	3 w	NR	−	0	3	0	0	0
2	8 w	NR	−	NR	2	+	0	8
3	4 w	−	−	0	3	0	0	15
4	8 d	NR	−	0	4	0	0	21
5	4 w	NR	−	NR	3	0	0	31
6	8 w	NR	−	0	2	0	0	51
7	5 w	−	−	15	3	0	0	0
8	7 w	NR	−	15	3	+	0	0
9	10 d	−	+	0	2	0	12	40
10	12 d	−	−	0	2	+ +	14	56
11	8 w	−	NR	NR	3	+	17	41
12	9 m	−	+	0	2	0	17	NR
13	8 w	NR	−	NR	3	+	21	56
14	2½ yr	NR	NR	22	3	+ +	30	60
15	8 d	−	−	13	2	+ +	36	47
16	2 w	+	+	NR	4	+ +	50	50
17	7 yr	NR	+	27	4	+ + +	58	68
18	4 d	NR	+	40	3	+ + +	75	90
19	4 w	NR	NR	NR	4	+ +	75	83
20	4 w	−	+	20	2	+ + +	83	93
21	3 d	−	+	0	4	+ +	89	93
22	3 d	NR	+	NR	3	+ + +	92	93
23	4 d	NR	+	NR	4	+ + +	93	95

*NR, not recorded, −, no evidence of stenosis, +, stenotic features present.
†0, not present, +, mild, + +, moderate, + + +, severe.

four cases was the electrocardiogram entirely normal.

SPECIAL INVESTIGATIONS
Only 10 patients underwent echocardiography (M mode in six cases and cross sectional in four). Seven of these studies correctly identified the pres-
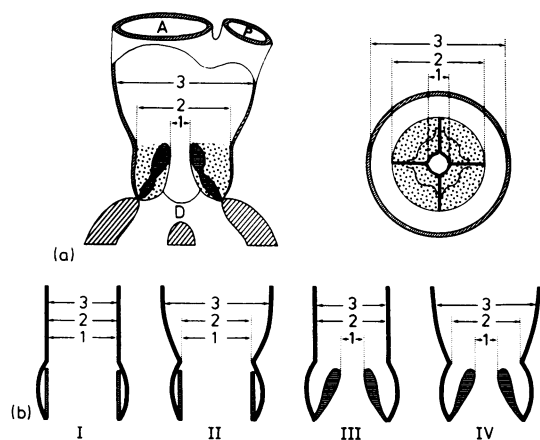

Fig. 1 *Diagrams showing (a) the measurement of absolute and relative stenosis (the truncus is seen in side view and from above) and (b) the concept of absolute and relative stenosis. A, aortic component of truncus; P, pulmonary component of truncus; D, ventricular septal defect; 1, maximum diameter of valve orifice; 2, diameter of valve ring; 3, maximum diameter of truncus; I, no stenosis; II, normal valve, relative stenosis only; III, stenosed valve, absolute stenosis only; IV, absolute and relative stenosis.*
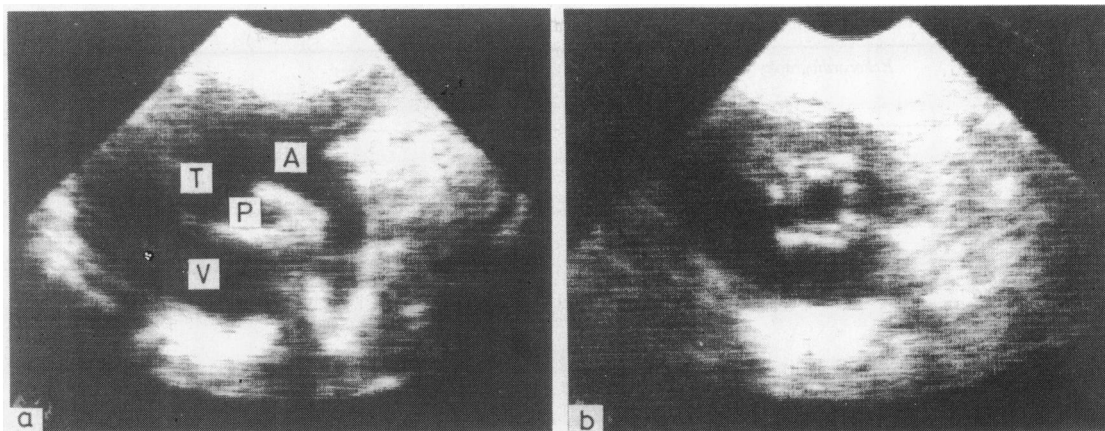
ence of a truncus arteriosus. In the M mode studies no mention was made of the appearances of the truncus valve, and the original recordings were not available for reassessment. In the four cases studied by cross sectional echocardiography stenosis of the truncus valve was predicted in the patient in case 16 (Fig. 2) but not in those in cases 7, 11, and 12 (Table).

Cardiac catheterisation and angiography was performed in 20 cases. In nine (45%) there were angiographic appearances suggestive of truncus valve stenosis, in that the cusps were thickened and had restricted movement (Fig. 3). Two of these also had angiographic evidence of regurgitation. In a further case there was isolated truncus valve regurgitation. Satisfactory pressure measurements across the truncus valve were available in 15 cases. In eight, there was no gradient; in the remaining seven the gradient ranged from 13 mm Hg to 40 mm Hg (mean 21·5 mm Hg) (Table).

MORPHOLOGY
Of the 23 specimens examined, in 15 there was a short pulmonary trunk which divided into right and left pulmonary arteries, in five the right and left pulmonary arteries arose separately from the left side of the truncus, and in three the right and left pulmonary arteries arose from opposite sides of the truncus or side by side from the posterior wall. These three varieties correspond to the morphological types 1, 2, and 3 according to the classification of Collett and Edwards.[15] There was one example of the so called

Fig. 2 *Cross sectional echocardiograms (a) in the suprasternal long axis view showing persistent truncus arteriosus and (b) in the parasternal short axis view in the same patient showing a stenosed quadricuspid truncus valve. A, aorta; P, pulmonary artery; T, truncus arteriosus; V, ventricle.*

type 4 truncus, in which the arteries to the lungs arose from the descending aorta, but this was excluded as this condition is now generally considered to be a form of pulmonary artery atresia with ventricular septal defect and not truncus arteriosus.

The truncus valve was bicuspid in seven, tricuspid in 10, and quadricuspid in six. Dysplasia, in the form of cusp thickening, nodularity, and commissural fusion, was present in 15 (65%) cases. In 10 of these the condition was severe enough to have produced appreciable valvar stenosis. Figures 4 and 5 show examples of stenotic and non-stenotic valves. Dysplasia was commoner in bicuspid and quadricuspid valves (eight of 13 (61·5%) cases) than in tricuspid valves (three of 10 (30%) cases).

Histological examination of dysplastic valves in two

Fig. 3 *Left ventricular angiogram showing stenosed and dysplastic truncus valve (arrowed).*

patients (cases 14 and 17) (Table) showed characteristic thickening by mucoid connective tissue.[3] Measurements, as described earlier, showed a degree of absolute stenosis varying from 12% to 93% in 15 cases. Isolated relative stenosis ranging from 8% to 51% was present in a further five cases (Table). No patients had branch pulmonary artery stenosis. In 10 patients there were associated cardiac malformations: right aortic arch, four cases; interrupted aortic arch, three; anomalous retro-oesophageal right subclavian artery, two; and bilateral superior caval veins, one.

Discussion

Abnormalities of the leaflets of the semilunar valve in truncus arteriosus are common and well described.[3 7 8] These abnormalities are associated with incompetence of the valve in 2–20% of cases.[7 8] By contrast, stenosis of the valve has been recognised infrequently, and very few well documented cases have been reported.[7 10–14] In their review of 100 cases of truncus arteriosus, however, Calder and colleagues noted pressure gradients across the truncus valve in 14 out of 45 patients, and truncus valve stenosis was present in eight out of 79 necropsy specimens.[8] Unfortunately, the criteria by which stenosis was judged to be present in the necropsy specimens were not stated. Assessment of stenosis of a semilunar valve, particularly when cusp dysplasia is present, is difficult in a fixed morphological specimen, and some authors have not attempted to give an opinion.[5] We have identified cases of truncus valve stenosis in pathological specimens by careful measurement of the maximum diameter of the valve orifice and relating this to the diameter of the truncus valve ring and the
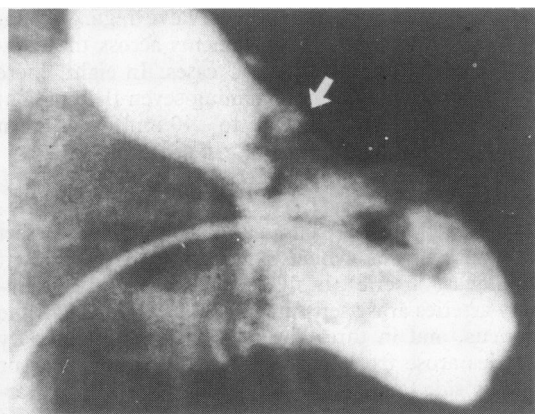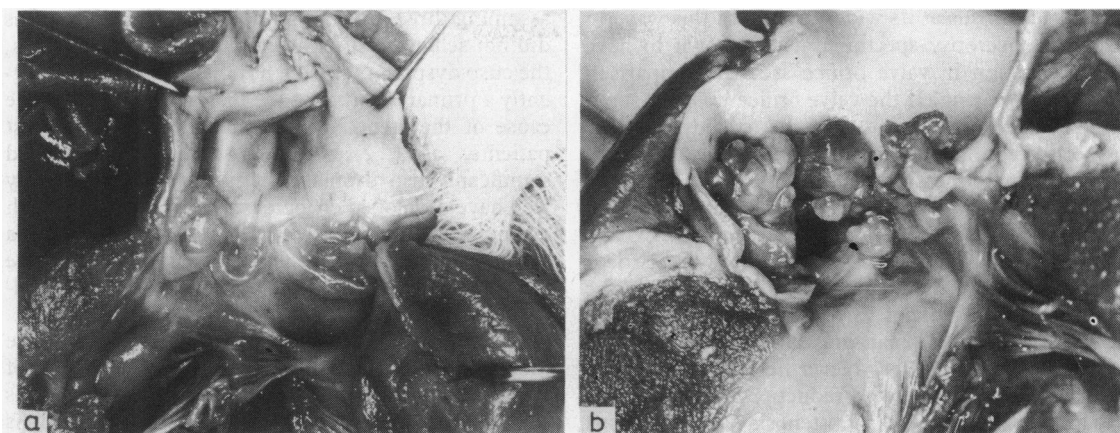
Fig. 4 *Morphological appearance of truncus valves opened across the valve ring: (a) quadricuspid valve without dysplasia or stenosis; and (b) stenosed and grossly dysplastic valve.*
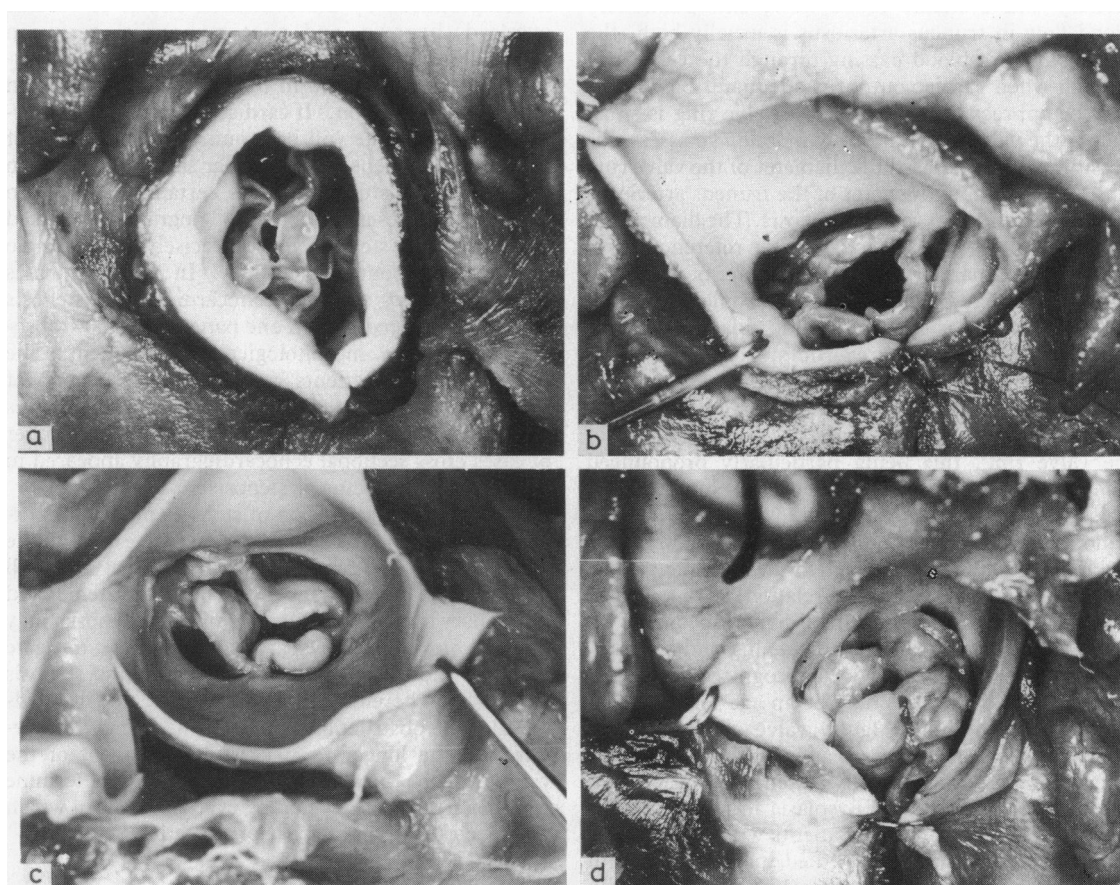

Fig. 5 *Morphological appearance of truncus valves viewed from above: (a) quadricuspid valve without stenosis or dysplasia; (b) bicuspid valve with moderate dysplasia and stenosis; (c) tricuspid valve with severe dysplasia and stenosis; and (d) quadricuspid valve with gross dysplasia and stenosis.*

truncus arteriosus at its widest point. In this way, 15 of the 23 necropsy specimens were shown to have some reduction in valve orifice area by comparison with the valve ring. If the valve orifice was compared with the maximum diameter of the truncus itself the proportion rose to 20 of 23. It is impossible to be sure to what extent measurements of this sort made on fixed morphological specimens reflect the situation in life, and for this reason minor degrees of stenosis calculated in this way should probably be disregarded. Similarly, it is impossible to define a "normal" size for the orifice of the truncus valve, and therefore we have deliberately avoided separating our cases into stenotic and non-stenotic groups. Nevertheless, one third of our cases (Table) showed a reduction in valve orifice area of ⩾50%, and clearly, stenosis of the truncus valve is more common than has previously been appreciated.

Since the truncus valve carries the combined pulmonary and systemic blood flow, and pulmonary blood flow in truncus arteriosus is usually high, the total volume of blood passing through the valve may be high when cardiac output is adequate. Blood flow may be appreciably limited if the valve ring is small even if the valve cusps are fully mobile. To assess this possibility we compared the diameter of the valve ring with the maximum diameter of the truncus arteriosus to obtain an index of relative stenosis. The diameter of the truncus is probably not the best reference point for such a comparison since the diameter of the vessel at this point may be influenced by turbulent flow distal to the valve as in poststenotic dilatation of the ascending aorta in valvar aortic stenosis. Nevertheless, since in most cases the aorta had been divided at the arch there was no practical alternative. Most specimens showed some degree of relative stenosis of the valve ring, this being particularly pronounced when there was also more than 50% absolute stenosis. This does perhaps suggest that when the valve is stenotic there is associated poststenotic dilatation and makes it difficult to make any definite statement about the size of the valve ring in relation to the cardiac output. Valvar stenosis was invariably associated with severe cusp dysplasia. Cusp thickening and nodularity, without stenosis, is a well recognised feature of truncus arteriosus. This may be an age related problem,[3][8] possibly due to faulty valve alignment, but Bharati *et al* found no association with age.[7] In our series there was a statistically significant inverse relation between the degree of dysplasia and the age of the patient at death. Of the 11 patients with moderate or severe dysplasia, nine (82%) died in the first month compared with five out of 12 (41·7%) patients with slight or no dysplasia (difference 40·3%; SE 18·3%). We found a positive relation between both stenosis and cusp dysplasia and an abnormal number of cusps,

assuming three to be normal (Table). The differences did not achieve statistical significance. In most cases, the cusp dysplasia, like the number of cusps, is apparently a primary feature of the malformation and is the cause of the stenosis. Nevertheless, the two oldest patients, aged 2½ and 7 years, both showed significant cusp dysplasia with stenosis, which may well have developed or increased with time. Although examples of neonatal and late cases of cusp dysplasia showed similar histological appearances, there may be two aetiological factors, both of which can cause valvar stenosis.

Although the haemodynamic data are incomplete the correlation between morphological evidence of stenosis and a pressure gradient across the valve is clearly poor. The presence of a pressure gradient is related not only to valvar stenosis but also to the flow across the valve at the time of measurement. In many patients with truncus arteriosus myocardial function is compromised by the combined effects of volume overload, hypoxia, and poor myocardial perfusion consequent on a low systemic arterial diastolic pressure.[16] This is reflected in the T wave changes often seen in this condition.[9] If cardiac output is low a pressure gradient alone will be an unreliable indicator of the severity of stenosis, and, as far as can be judged in a retrospective study, this was certainly the case in many of the present patients. Angiography identified the cases with stenosis more precisely but gave no information about the severity. In the four cases studied by cross sectional echocardiography stenosis was correctly predicted in one patient with 50% absolute stenosis on morphological measurement. The remaining three patients had only minor degrees of absolute stenosis (maximum 17%), and the valves were not identified as stenotic. In this limited number of cases cross sectional echocardiography appeared to be a reliable predictor of stenosis.

Preoperative recognition of stenosis may be important since after repair the truncus valve lies in the aortic position. Although flow through the valve will have been reduced by the separation of systemic and pulmonary circulation any significant stenosis may further compromise myocardial function and mitigate against a successful outcome. Recognition of stenosis might lead therefore to measures directed towards relief of the obstruction as part of the primary procedure. In the longer term careful assessment of the valve will be necessary at intervals to determine whether stenosis is developing even in valves in which no stenosis was apparent at presentation.

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